ESID Online Registry

User Manual

provided by:

www.uniklinik-freiburg.de/cci.html

Version 1.8

2015-07-13

Table of contents

1 Introduction 3
2 Working with the Database System 3
2.1 General principles 4
2.1.1 Two types of “unknown” 5
2.2 Accessing the system 6
2.3 The main page (patient list) 7
2.4 Adding a new patient 8
2.5 Veryifing an imported patient 8
2.6 The Patient tabs 11
2.6.1 Patient 12
2.6.2 Consent (ICF) 12
2.6.3 PID Diagnosis 13
2.6.4 Current status 15
2.6.5 Ig replacement 16
2.7 Deceased patients 18
2.7.1 ICD10 coding death causes 18
2.8 Follow-up documentation 20
2.8.1 Changing the PID diagnosis 21
2.8.2 Loading previous data for Ig replacement 23
2.9 Personalized version 24
2.9.1 Patient list with names 24
2.9.1.1 Improving the loading time 24
2.9.1.2 Downloading the patient list with names 25
2.9.1.3 Sorting and filters 25
2.9.2 Create a new patient 25
2.9.3 Edit personal information 27
2.9.4 Add personal information to existing patient 27
2.10 Account settings 28
3 Contacting the Registry administrator 29
4 Registry FAQ 30

1 Introduction

Welcome to the ESID Online Registry for Primary Immunodeficiencies. This web-based database system allows you to enter and access patient data in a secure decentralized way through the internet via your own browser. This manual includes a detailed guide to the documentation process.

All pages in the system include detailed descriptions (tool tips and other explanations). Therefore, most of the features should be self-explanatory. With this manual, we mainly want to describe the general documentation process and those features which are probably not completely “intuitive”.

The ESID Registry project is managed at the Center for Chronic Immunodeficiency at University Medical Center Freiburg.

Current project leader: Dr. Nizar Mahlaoui, Chairman ESID Registry Working Party
Project coordination: Dr. Gerhard Kindle, Benjamin Gathmann

2 Working with the Database System

Before we start this tour, we would like to remind you that you are working with real patient data. Therefore, please observe the following user principles:

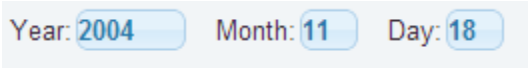
- every user of the system (= every user name) corresponds to one person only.
- never log into the system with someone else's user name.
- close the session by logging out after finishing your work.
- keep your password in a secure place
- use a secure password
- change your password regularly

2.1 General principles

- Please make a selection **for every field** – this is necessary in all forms of the Registry (if not stated otherwise).
- To save the data, you **must** click the “Create” or “Save” button below every form.
- Some centres use a **personalized version** of the system, i.e. they can work with patient names. The additional features of this version are described in 2.9
- Buttons with a question mark: contain definitions for all fields where necessary.
- The system contains “classic” drop down menus as well as searchable auto-complete fields. The latter are marked by a magnifier like this:

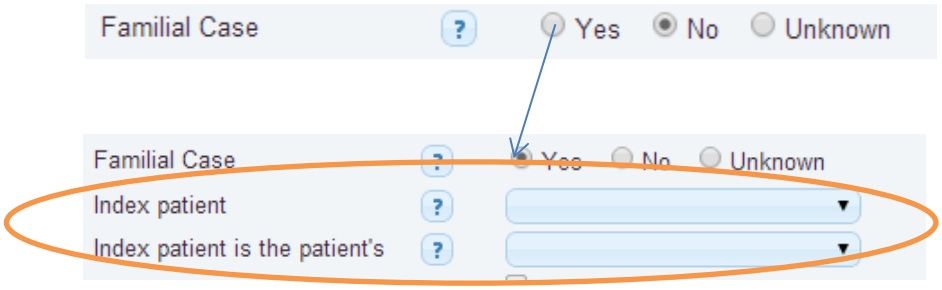
  To use this type of field
  1. Enter two or more characters
  2. Wait until a list of matching entries is displayed and select one of these
- Date fields:
  - Fields that ask for a complete date contain a date picker that appears upon selection of the field:

  - Date fields that are split into three fields:

    Here, it is possible to leave month and day open. Please only enter the date as precisely as known. Do not use dates like 2000-1-1 or 2000-15-6 to signal that month and day are unknown. This only leads to flaws in data quality.

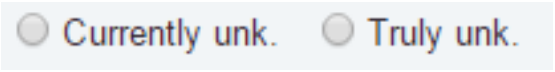
- Some fields only appear after you have selected a specific field, e.g.:



- If you have not completed a field or entered inconsistent data, messages are displayed on top of the page:

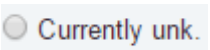
2.1.1 Two types of "unknown"

Since version 1.6, the ESID Registry offers two fields to indicate that information for an item is not available:

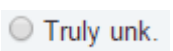


These have been introduced to enable the documenting centres to differentiate between items that they currently don't have at hand but may be able to enter it later after further investigation, and items where data is really not available not matter how hard they look. Before this was introduced, there was only one option "unknown" which did not offer this possibility. So, please note the following

Definitions:



This means that you do not have the data at hand at the moment, but if you ask the treating physician or go trough the patient files in detail, you may find and enter it later.



This means that data is really not available, and in effect you should waste no more time looking for it.

All previously entered "unknown" items have been converted to "currently unknown" when switching to version 1.6 (March 13, 2015).

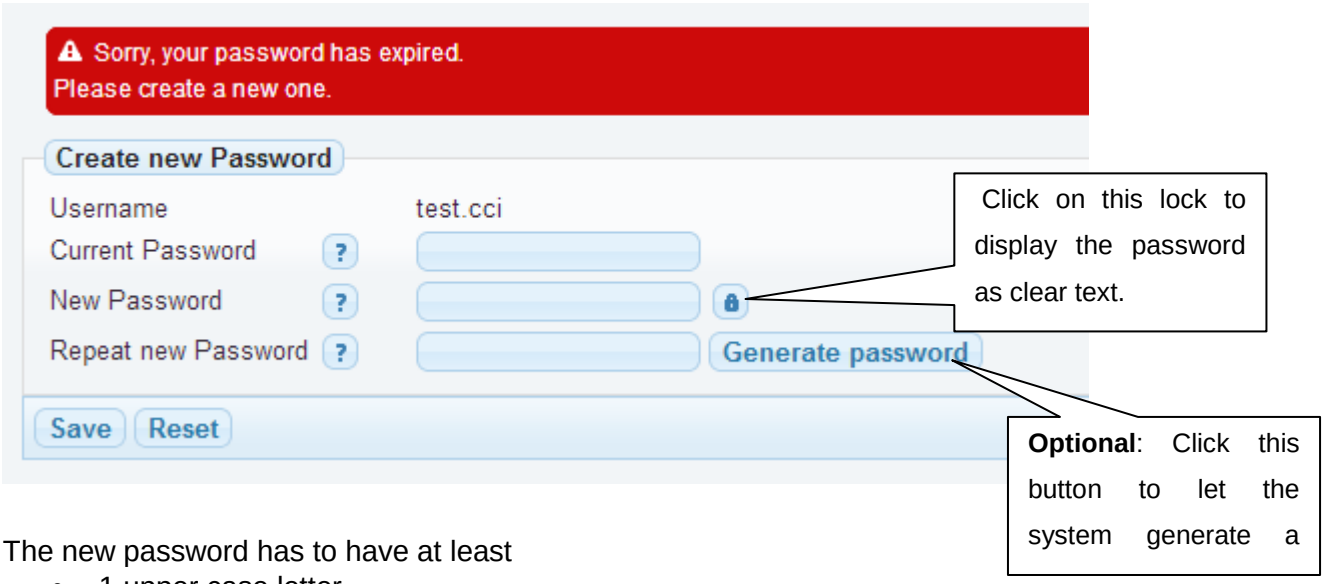
2.2 Accessing the system

Enter the following URL in your browser:

https://cci-a1.ukl.uni-freiburg.de/EERS

Enter the login credentials you received from the ESID Registry team.

When you log in for the first time, you will be asked to change your password:



The new password has to have at least

- 1 upper case letter,
- 1 lower case letter,
- 1 digit,
- 1 special character of this list: +-_,:;=?!#

and has to be between 6 and 16 characters long.

In case you have trouble logging in, you find our contact details in the HELP menu (top right).

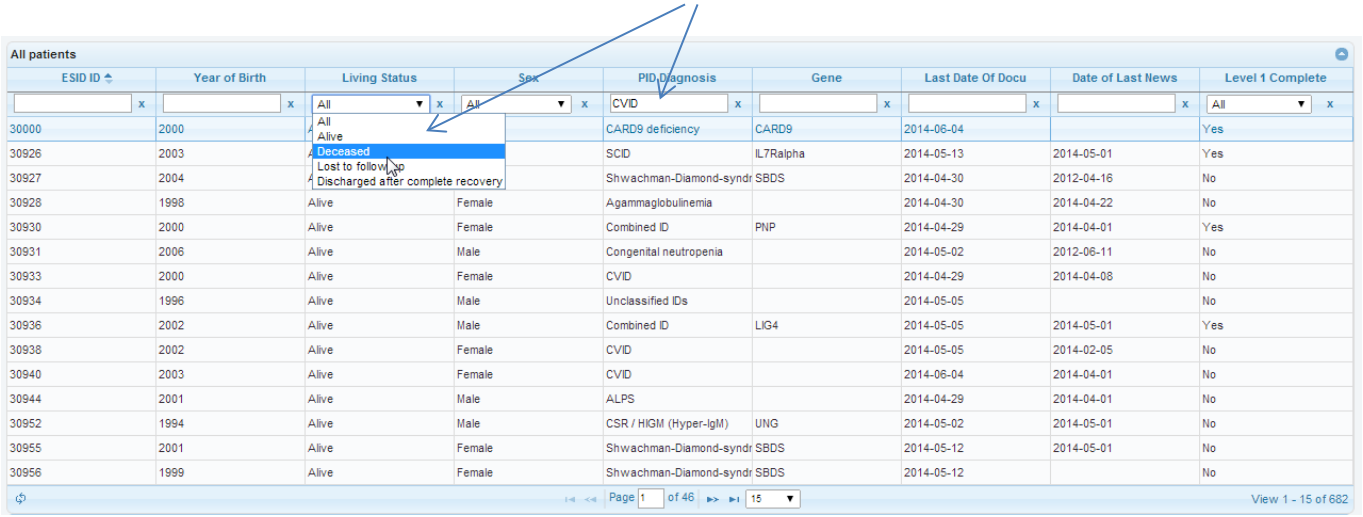
2.3 The main page (patient list)

After logging in, the system shows a list of your patients. This is the view that the system also returns to if you click “Select patient” in the top left corner.

The following columns are displayed:

- ESID ID: The unique patient ID
- Year of Birth
- Living Status (alive, deceased…)
- Sex
- PID Diagnosis
- Gene
- Last date of docu: The date of the last documentation timepoint
- Date of last news: The date that the most recent documentation is based on (as entered in form “Current status”)
- Level 1 Complete: “Yes” if level 1 was completed at the last date of docu

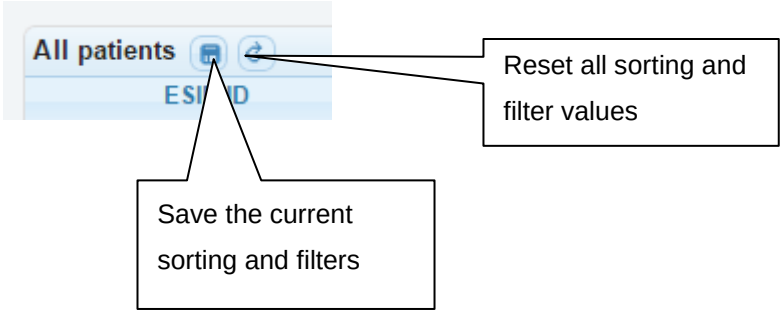
Each column can be used to **sort or filter** the list, e.g.:



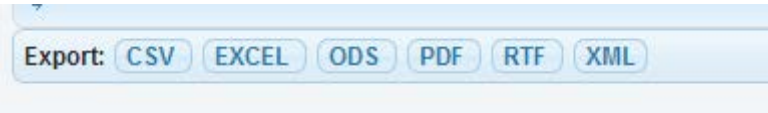
ESID ID	Year of Birth	Living Status	Sex	PID Diagnosis	Gene	Last Date Of Docu	Date of Last News	Level 1 Complete
30000	2000			CARD9 deficiency	CARD9	2014-06-04		Yes
30926	2003			SCID	IL7Ralpha	2014-05-13	2014-05-01	Yes
30927	2004			Shwachman-Diamond-syndr	SBDS	2014-04-30	2012-04-16	No
30928	1998	Alive	Female	Agammaglobulinemia		2014-04-30	2014-04-22	No
30930	2000	Alive	Female	Combined ID	PNP	2014-04-29	2014-04-01	Yes
30931	2006	Alive	Male	Congenital neutropenia		2014-05-02	2012-06-11	No
30933	2000	Alive	Female	CVID		2014-04-29	2014-04-08	No
30934	1996	Alive	Male	Unclassified IDs		2014-05-05		No
30936	2002	Alive	Male	Combined ID	LIG4	2014-05-05	2014-05-01	Yes
30938	2002	Alive	Female	CVID		2014-05-05	2014-02-05	No
30940	2003	Alive	Female	CVID		2014-06-04	2014-04-01	No
30944	2001	Alive	Male	ALPS		2014-04-29	2014-04-01	No
30952	1994	Alive	Male	CSR / HIGM (Hyper-IgM)	UNG	2014-05-02	2014-05-01	No
30955	2001	Alive	Female	Shwachman-Diamond-syndr	SBDS	2014-05-12	2014-05-01	No
30956	1999	Alive	Female	Shwachman-Diamond-syndr	SBDS	2014-05-12		No

Saving your sorting and filters:

If you would like to save your current filters, so that the list is loaded the same way the next time you call it, you can do so using the buttons on the left top side of the list:



The list can be exported in different formats using the buttons displayed below the list

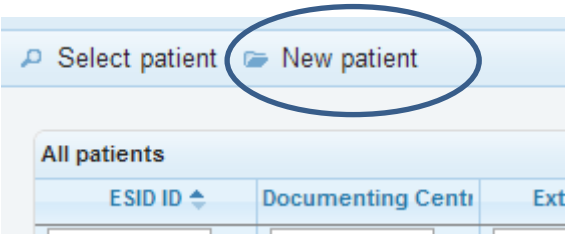


By default, the list shows 15 entries per page. You can set a personal default value of up to 100 entries in your account settings (2.10).

For users who enter data for more than one center, an additional column "Documenting Centre" is visible in the patient list.

2.4 Adding a new patient

In order to add a new patient, select "New patient" on the top left of the page:

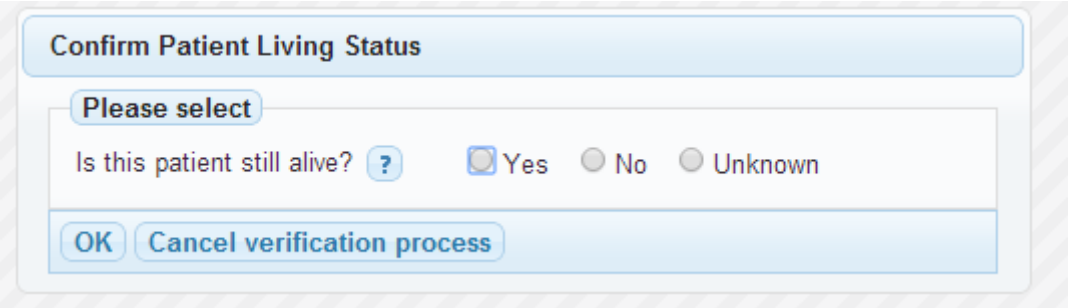


The following page shows up. Please enter the details for the patient.
Explanations for the fields are available via the "?" buttons.

After you have successfully created the patient entry, the standard patient tabs are shown (see 2.6)

2.5 Veryifing an imported patient

In order to verify an existing entry, click on the entry in the list. You are then asked to confirm that the patient is still alive (This does not apply for deceased patient entries transferred from another registry).

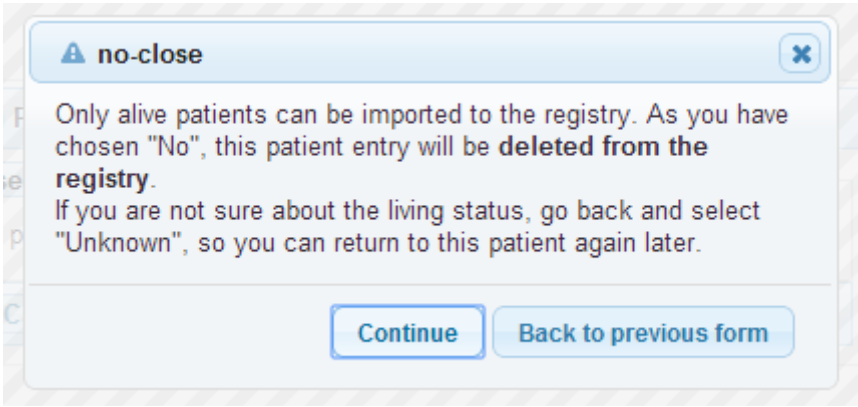


If the patient was still alive at the last time you heard of him, select "Yes". You can enter the time point of last news later on in the "Current status" form (2.6.2). This also means that: If the last time you saw him was a long time ago (e.g. two or three years), but your center still follows this patient, select "Yes".

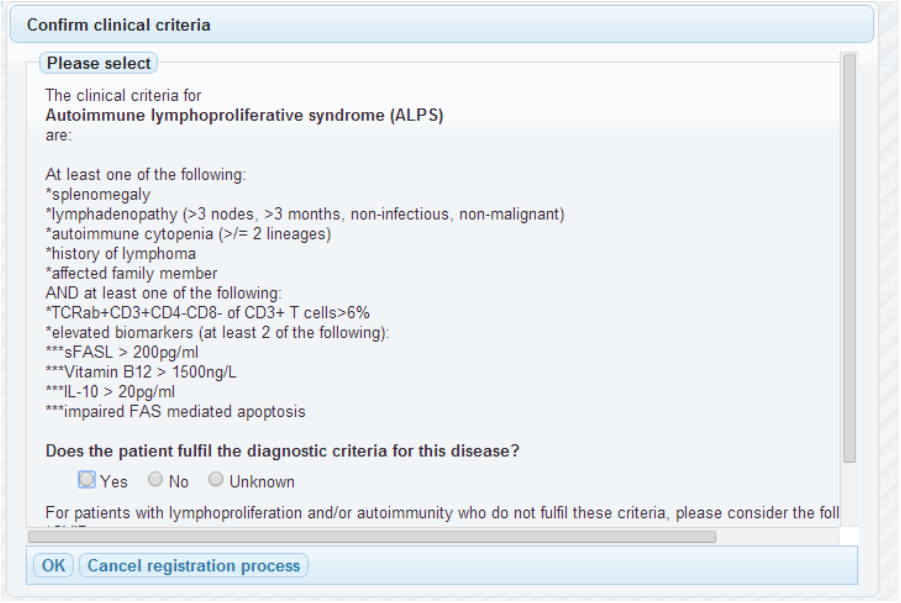
If the last time you saw him was a long time ago (e.g. more than three years) and your center does NOT follow him anymore, select "No". This is because you should only register patients currently followed by your center.

If you know that the patient is now followed by another center, and this is an ESID Registry Documenting Center, please send us an email with the patient ID. We can arrange a transfer of this entry to the other center then.

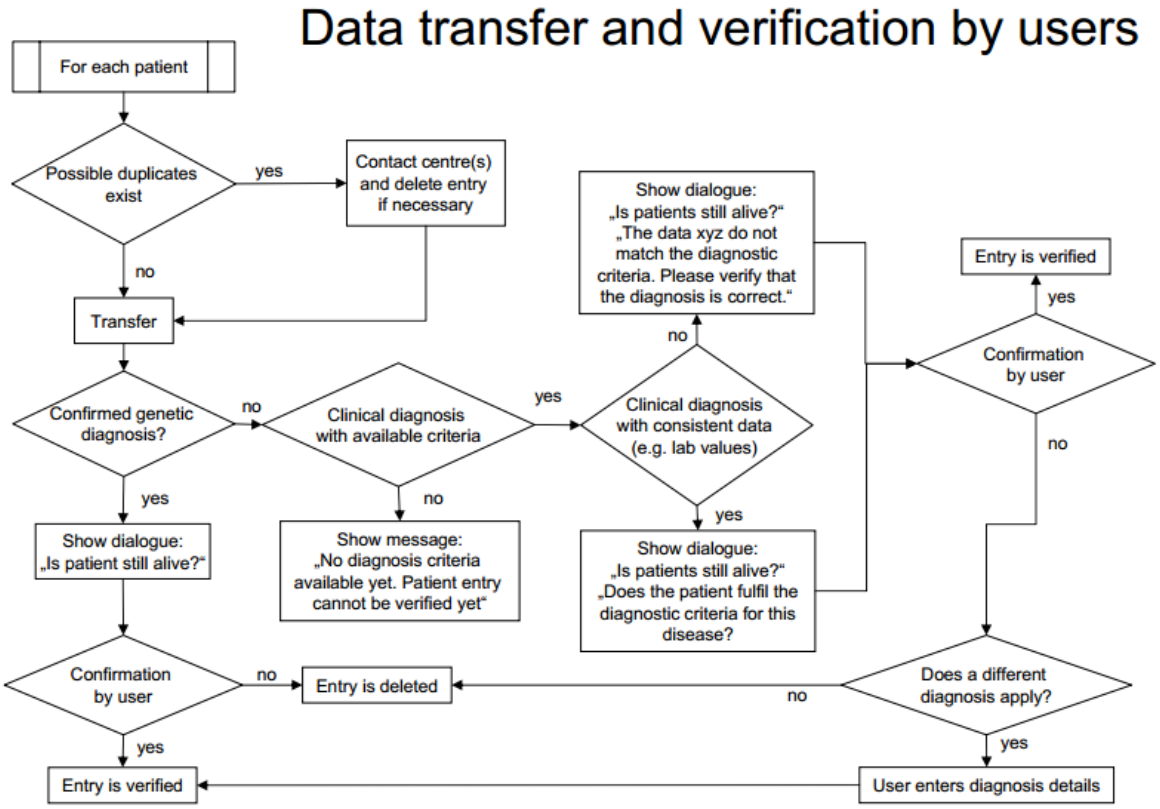
If you select “No”, the entry will be deleted from the system. This is because the system is not intended for retrospective documentation of deceased patients.
The system asks you for confirmation before the entry is deleted:


no-close
Only alive patients can be imported to the registry. As you have chosen "No", this patient entry will be deleted from the registry.
If you are not sure about the living status, go back and select "Unknown", so you can return to this patient again later.
Continue
Back to previous form

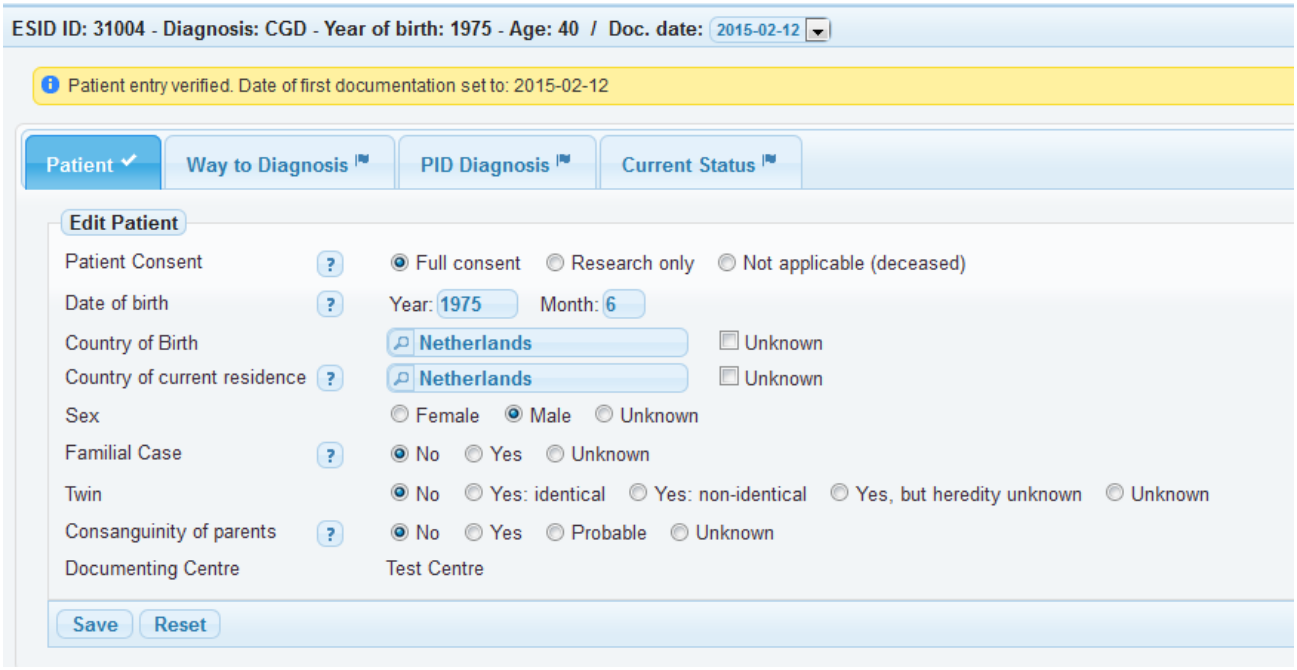
Next, depending on the type of diagnosis, the system asks you to confirm or change the diagnosis, e.g.:


Confirm clinical criteria
Please select
The clinical criteria for
Autoimmune lymphoproliferative syndrome (ALPS)
are:
At least one of the following:
*splenomegaly
*lymphadenopathy (>3 nodes, >3 months, non-infectious, non-malignant)
*autoimmune cytopenia (>/= 2 lineages)
*history of lymphoma
*affected family member
AND at least one of the following:
*TCRab+CD3+CD4-CD8- of CD3+ T cells>6%
*elevated biomarkers (at least 2 of the following):
***sFASL > 200pg/ml
***Vitamin B12 > 1500ng/L
***IL-10 > 20pg/ml
***impaired FAS mediated apoptosis
Does the patient fulfil the diagnostic criteria for this disease?
Yes
No
Unknown
For patients with lymphoproliferation and/or autoimmunity who do not fulfil these criteria, please consider the foll
OK
Cancel registration process

The underlying process is depicted here:



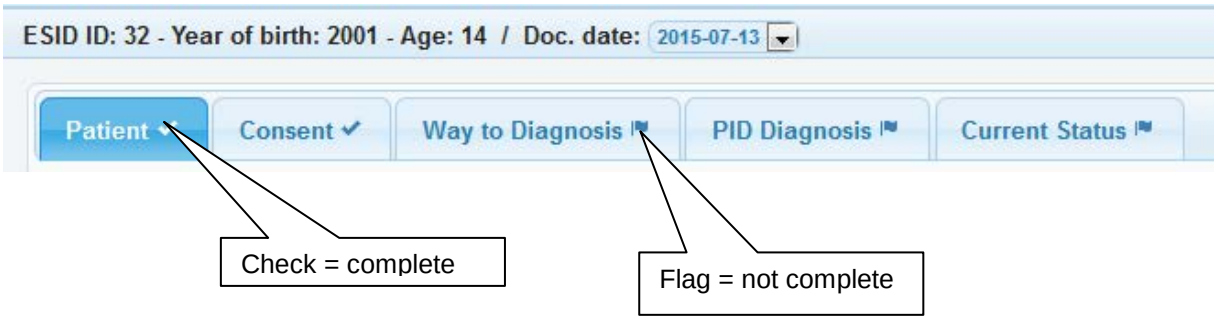
If you have successfully verified the entry, the patient tabs are shown for further editing (see 2.6).



2.6 The Patient tabs

The patient tabs are the standard view for a patient.

There is a header row that shows the patient's unique ID, the year of birth and current age, as well as the currently selected Documentation date:



The tabs themselves carry checks and flags that signal whether the form has already been completed.

If you are editing a patient that has been imported, some forms may already contain data, but the forms are not marked as complete, because selections are missing for some items.

In order to complete a patient, please select every tab marked by a flag and enter the necessary data.

The Documentation date which is shown on the top right cannot be edited manually. It is the time point when the patient was created (or verified, see 2.5) and inserted automatically by the system.

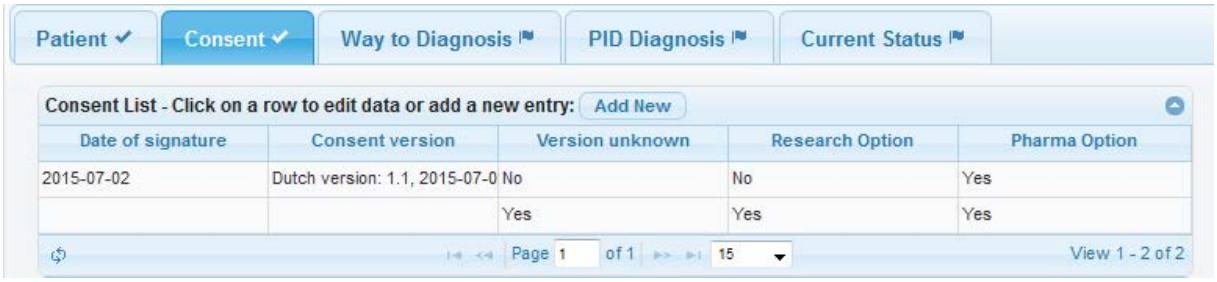
Some of the tabs (forms) are described in detail in the following chapters.

2.6.1 Patient

If you select “Yes” for familial case, you can select the ID of the index patient for this family (if he/she has been registered). The **index patient** is the first patient diagnosed with this immunodeficiency in this family. If you link several patients in this way, you are able to switch between patients belonging to this family by clicking on “show family list”:

2.6.2 Consent (ICF)

Since version 1.8, the system requests detailed information on the ICF (informed consent form) that the patient has signed. Note that you can add several consent forms for a patient, like here:



For all registered patients until the end of version 1.7 (2015-07-13), the ICF version has been set to "unknown", so that datasets are still evaluated to complete.

Please note that you can select from all consent forms that have ever been in use at your centre. The consent forms are each distinct and follow this pattern:

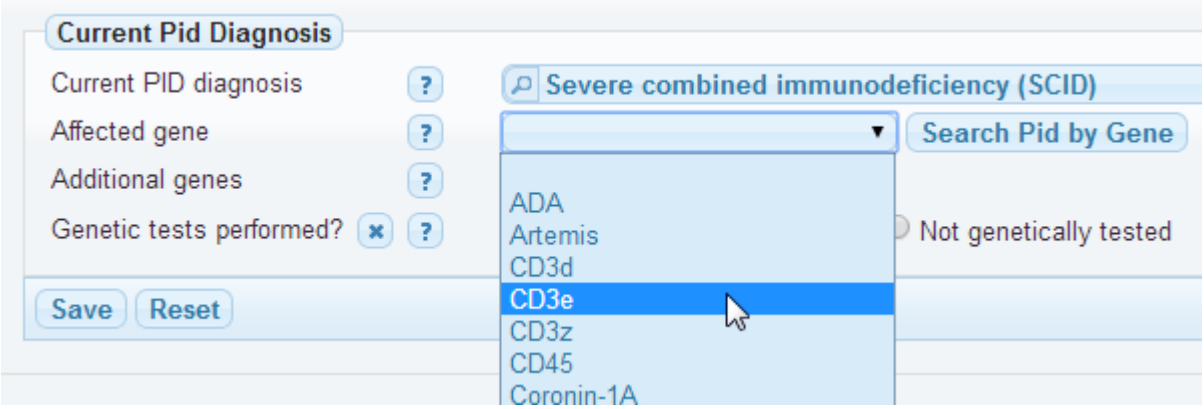
Language **-** Version number **-** Version date **-** ethics committee (EC) that approved this form

If a version is missing, please send an email to us, we will add it for you.

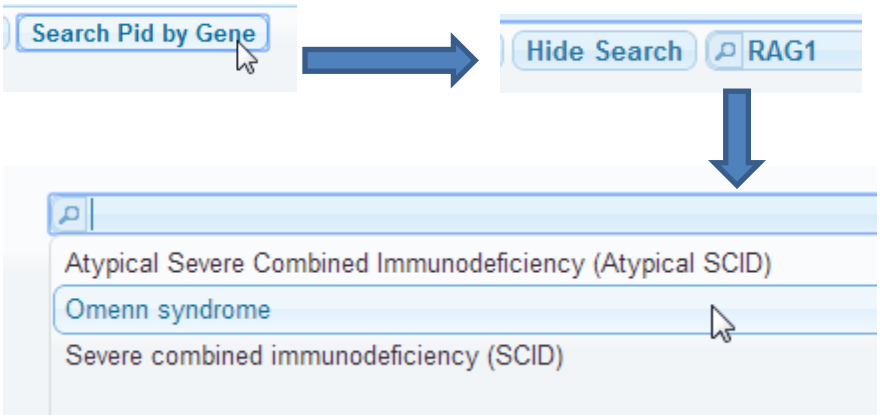
2.6.3 PID Diagnosis

In the PID Diagnosis form, it is possible to enter a diagnosis with known or unknown genetic defect:

- Known genetic defect:
  You can either select a clinical diagnosis (“Current PID diagnosis”), and the possible genes for this diagnosis are displayed:



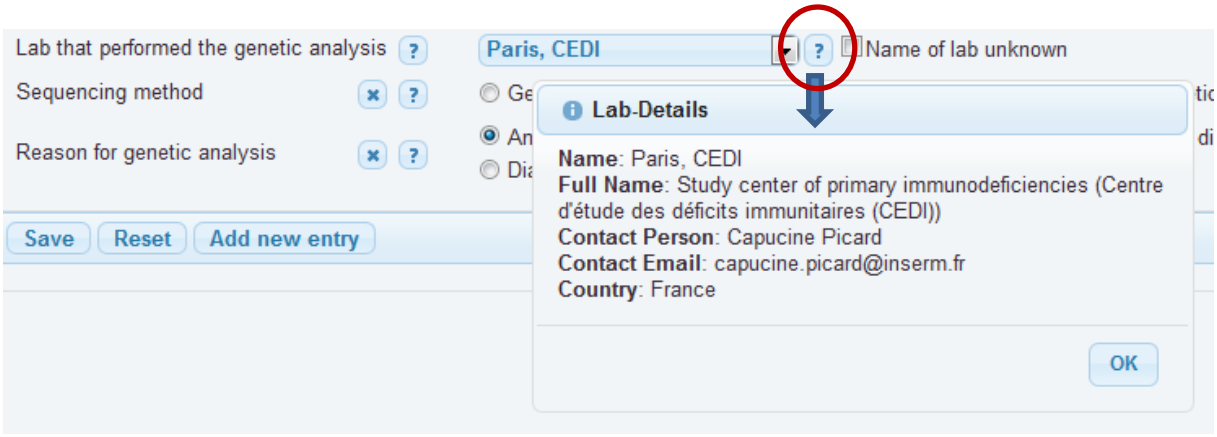
  OR you can first enter the gene and then select a suitable PID diagnosis:



- Unknown genetic defect:
  Use the “Current PID Diagnosis” field to select a PID diagnosis. Next, you will be asked to confirm the clinical diagnosis criteria (as described in the verification process (2.5).

Genetic laboratories:

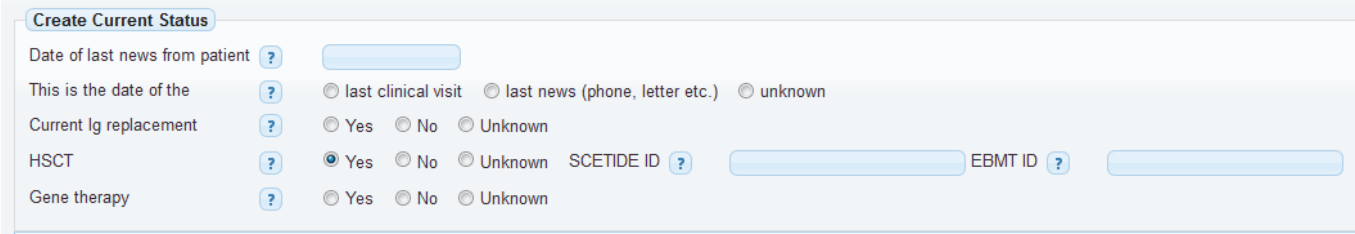
There is a field that asks for the name of the “Lab that performed the genetic analysis”. You can select one of the names from the list and see the details for the lab by clicking on the question mark if you want to make sure that this entry is correct:



2.6.4 Current status

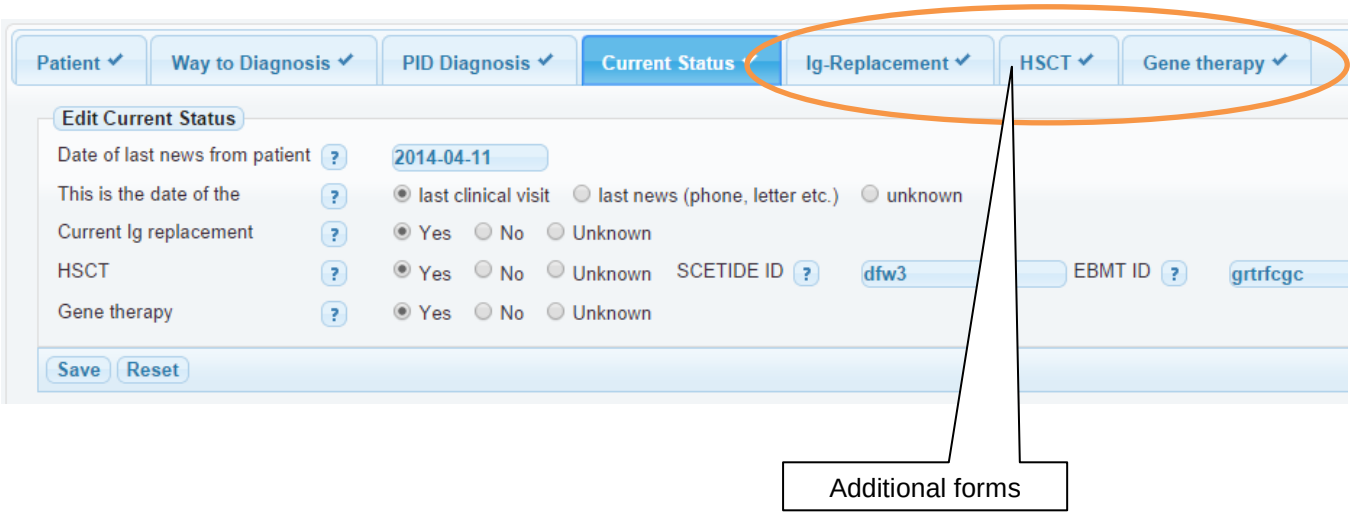
Patient, Way to Diagnosis and PID Diagnosis only have to be completed once (at the first documentation = baseline documentation). The Current Status form has to be completed at every documentation timepoint.

This is what the Current Status form looks like at baseline:



Please note the following:

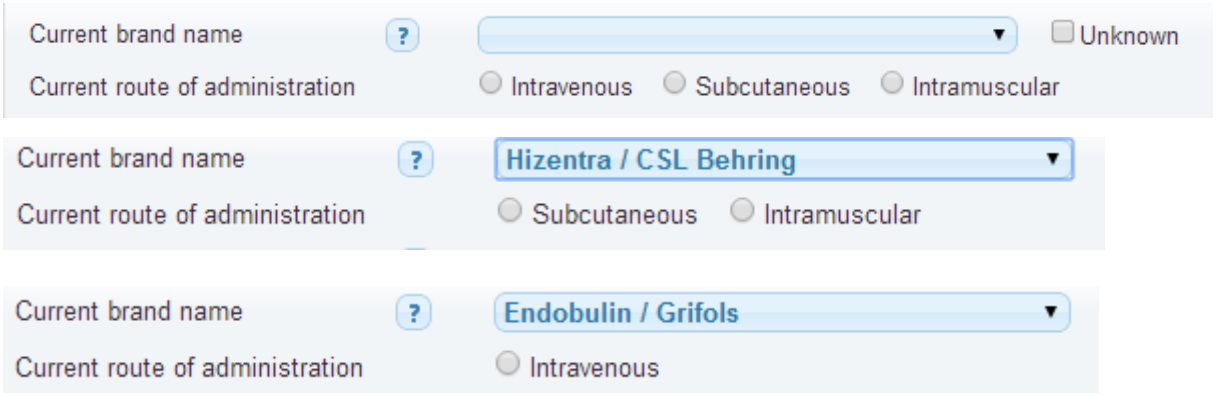
- You must enter a “date of last visit” of the patient at your centre or the “date of last news” from the patient. This is important to know which timepoint the reported data refers to.
- If you select “Ig replacement”, an additional tab (form) appears.
- The same is valid for HSCT and Gene therapy.



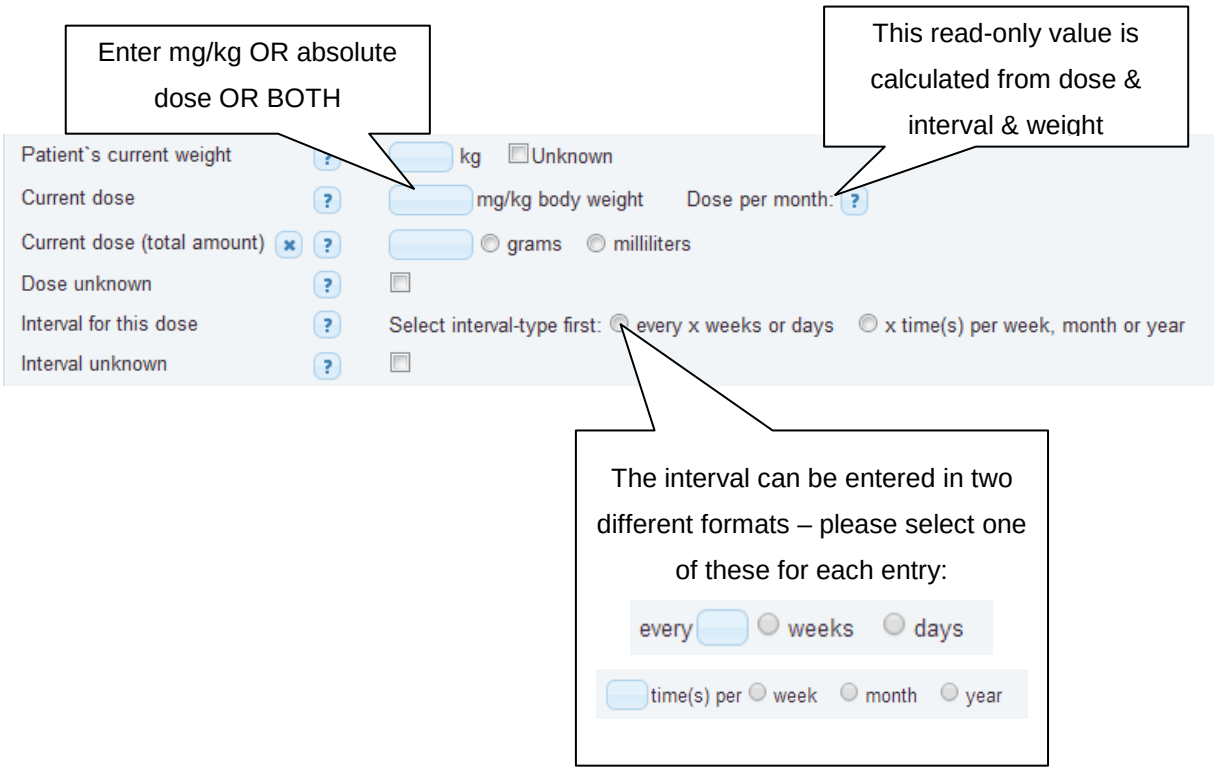
2.6.5 Ig replacement

The form for immunoglobulin replacement includes many automatic checks as well as calculations in order to ensure that data is entered complete and correct.
Some of these features are described here:

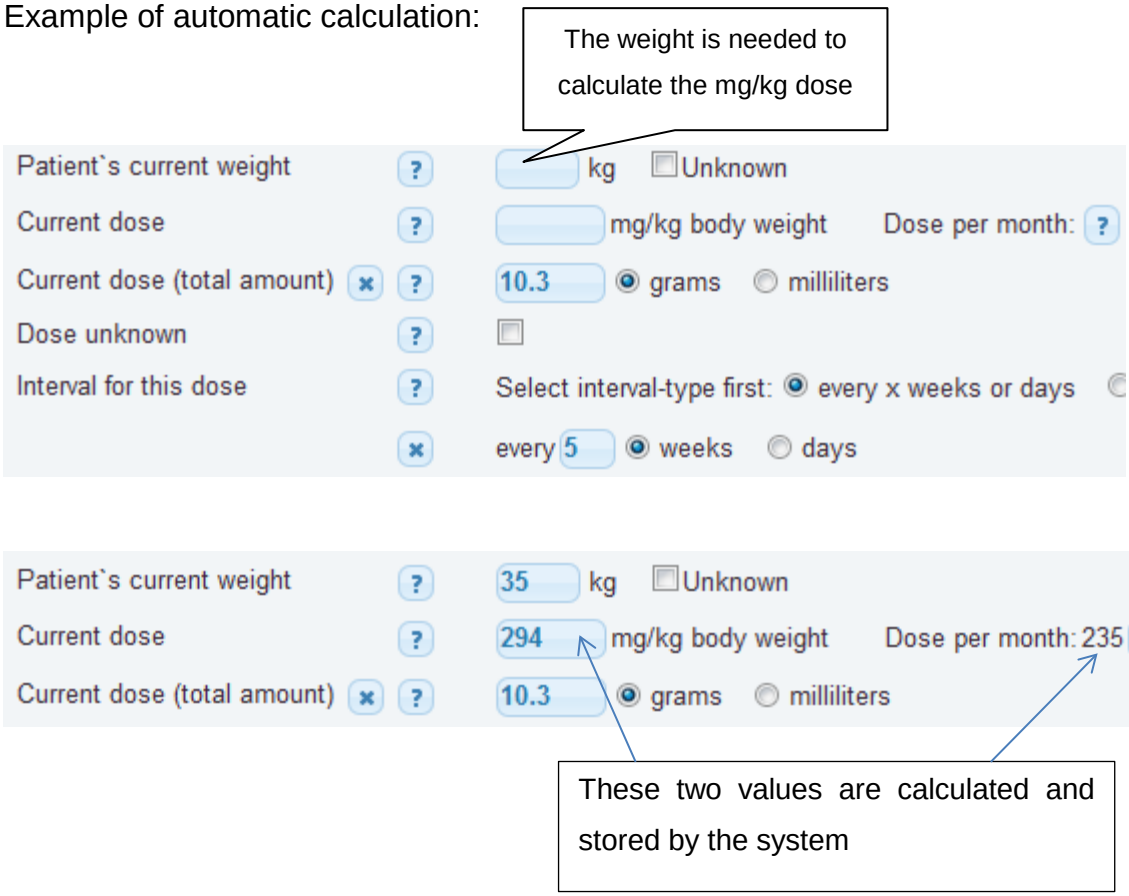
- Only appropriate routes are shown for each brand:



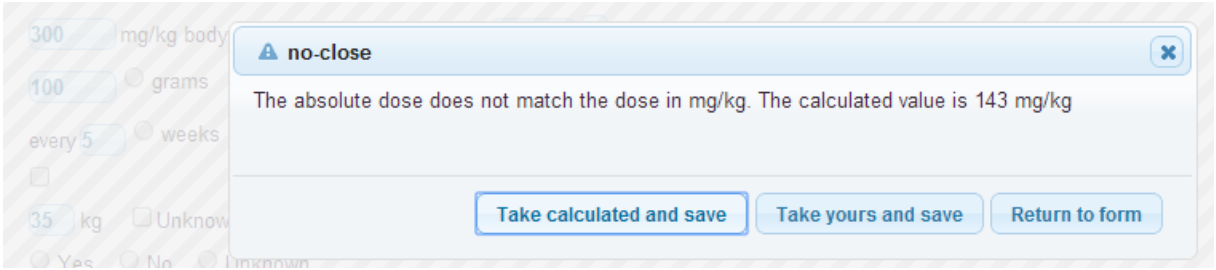
- Automatic calculation of dose:



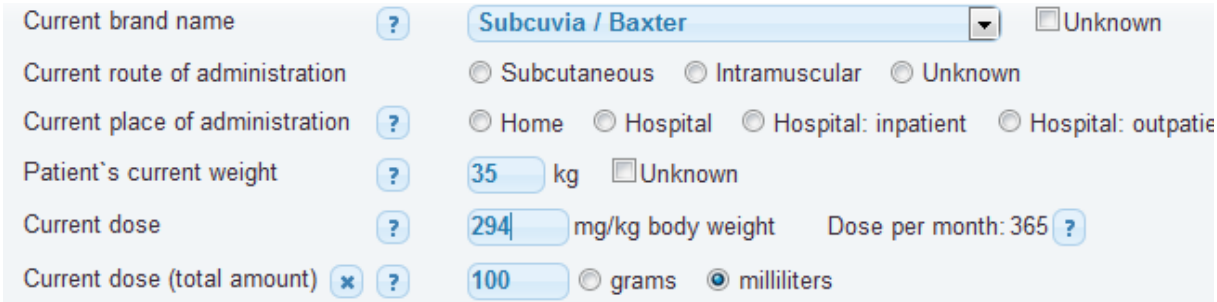
Example of automatic calculation:



If you enter BOTH mg/kg dose and absolute dose, the system calculcates whether the two match. If not, a dialog box is shown as follows:


no-close
The absolute dose does not match the dose in mg/kg. The calculated value is 143 mg/kg
Take calculated and save
Take yours and save
Return to form

If you enter the dose in milliliters, the brand name is needed for the calculation, because each brand has a specific IgG concentration:



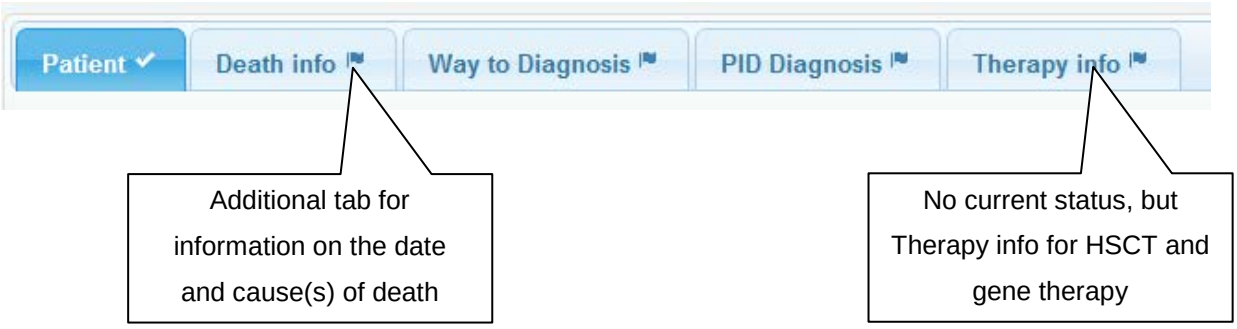
2.7 Deceased patients

If you are

a) working on an imported entry of a deceased patient or

b) have selected “Death before initial registration” for a new patient,

the Patient tabs look slightly different:

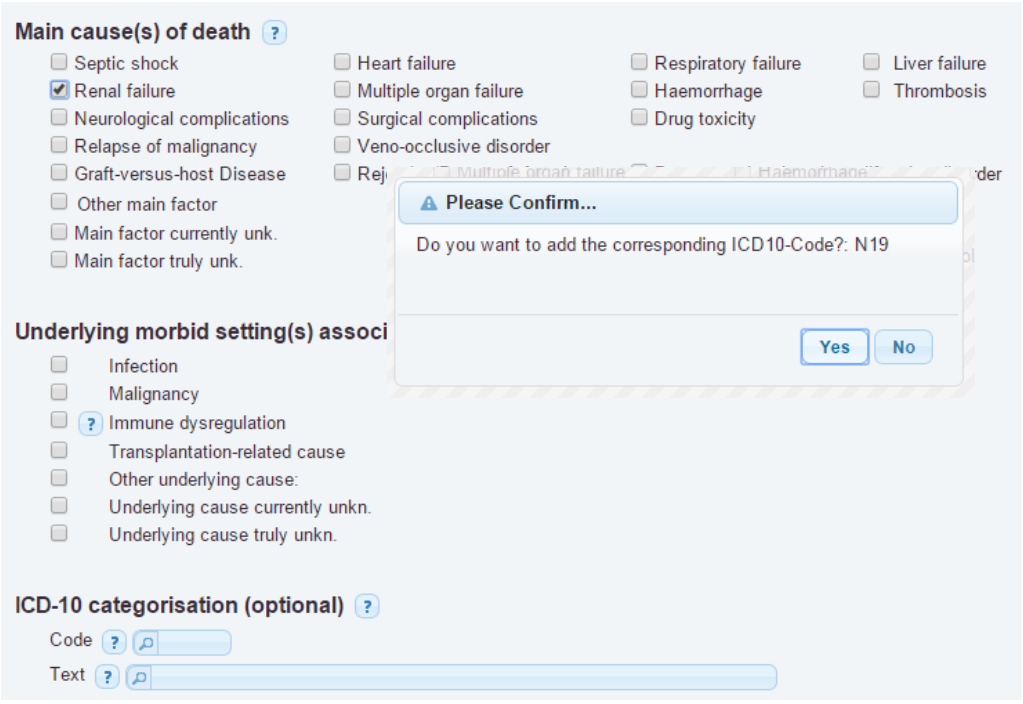

Patient
Death info
Way to Diagnosis
PID Diagnosis
Therapy info
Additional tab for information on the date and cause(s) of death
No current status, but Therapy info for HSCT and gene therapy

Please note that no follow-ups can be entered for deceased patients. Previous entries for Ig replacement remain stored. Data on HSCT and gene therapy can still be entered.

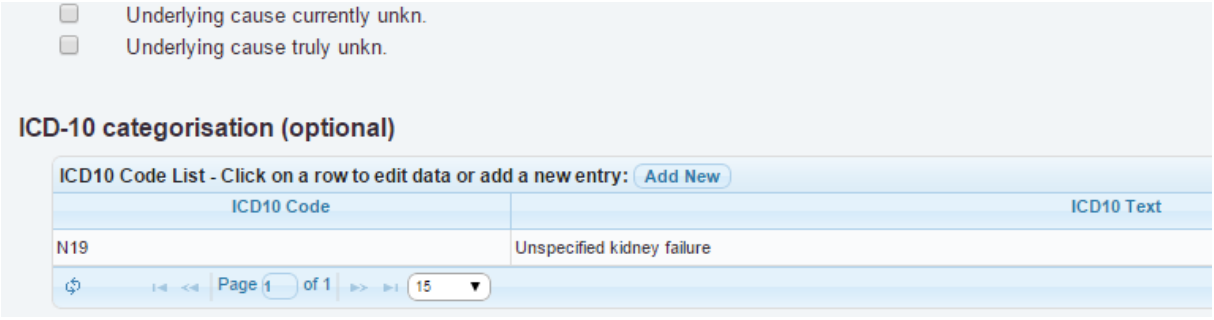
2.7.1 ICD10 coding death causes

The “Death info” tab contains an additional section for ICD10 codes. This section is – in contrast to all other items in the Registry – optional. It was added because some centres need this categorization for internal purposes.

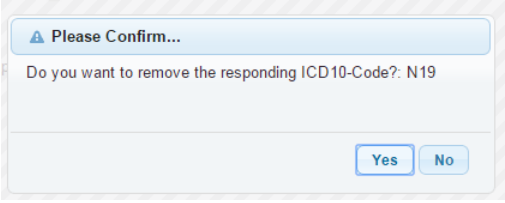
In order to simplify the adding of ICD10 codes, corresponding codes for the “Main cause of death” are suggested by the system when one of these options is selected.



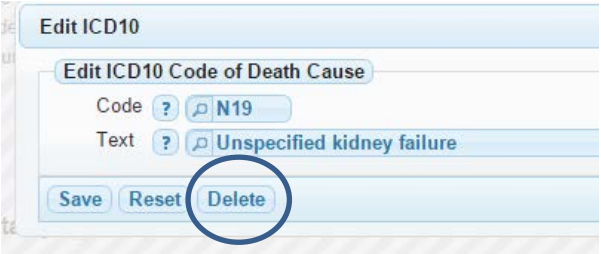
If confirmed, the respective entry is added to the ICD10 section:



If an option is de-selected, the system will ask if the corresponding ICD10 entry shall be removed:



Of course, you can also add and edit ICD10 codes manually. You can also delete them by selecting an entry and then click the “Delete” button:



2.8 Follow-up documentation

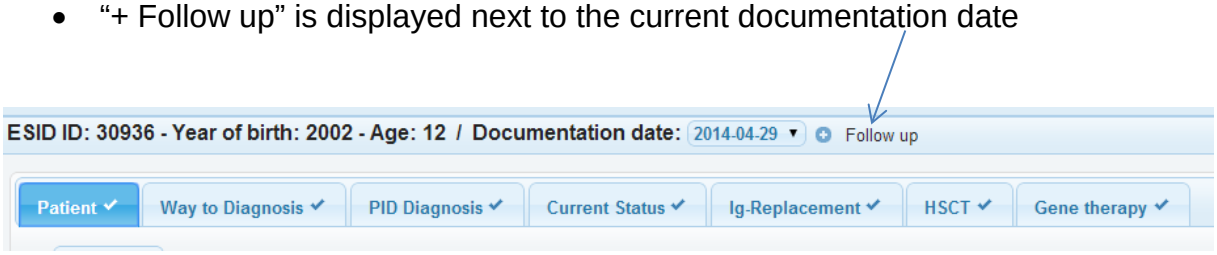
If you have completed a documentation timepoint for a patient, it is possible to add a follow-up documentation. The system will start sending email reminders if a patient has not been updated for more than one year (365 days).

In any case, you can enter follow-ups as often as you like (but only one per day).

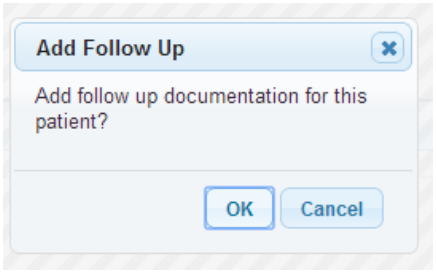
Please note though that only one documentation timepoint is possible per day. This also implies that you cannot enter baseline data and follow-up data on the same day, because the documentation date is automatically generated and stored by the system.

This is an example for a complete documentation timepoint:

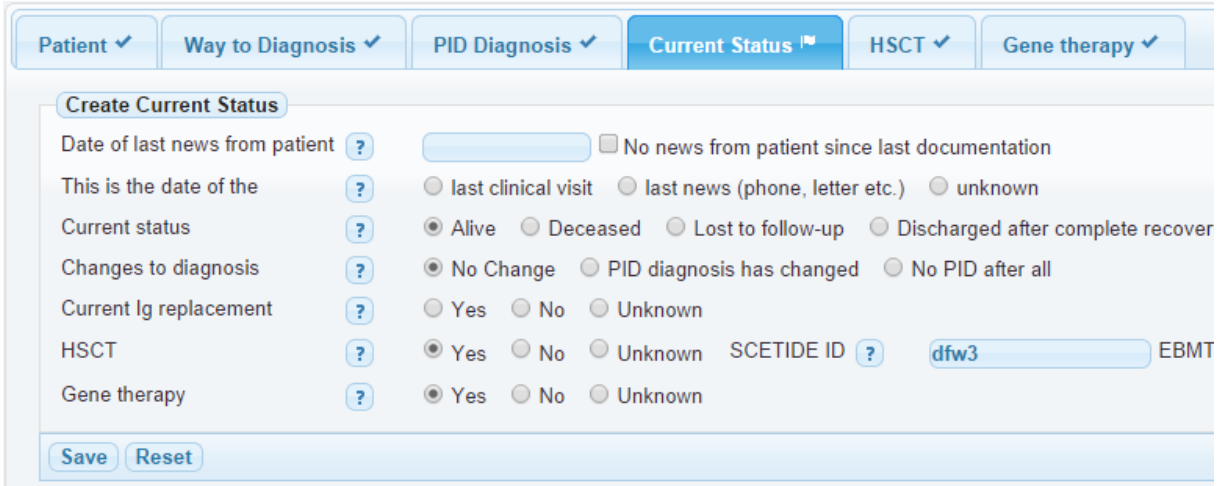
- All tabs have a check sign
- "+ Follow up" is displayed next to the current documentation date



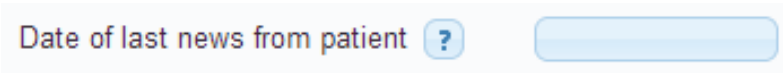
The following dialog box is shown:



Next, the patient tabs are shown, and the Current Status form is selected:

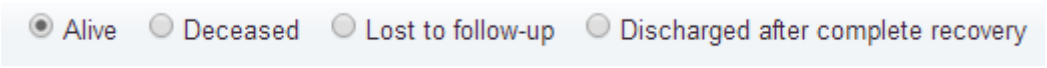


As you can see, all other tabs do NOT have to be documented again, except for Ig replacement. Only the following items require documentation:



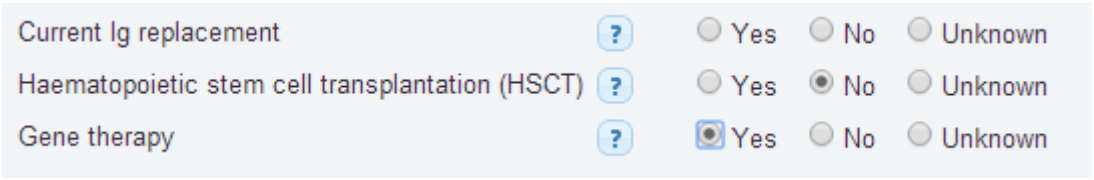
(or alternatively the date of last news from the patient)

Current status (the previous entry is selected):



Changes to diagnosis (No change is pre-selected)

Therapy items:

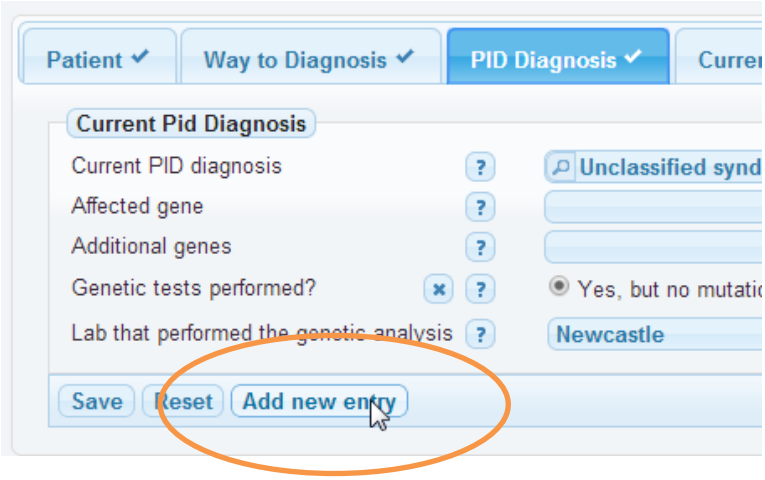


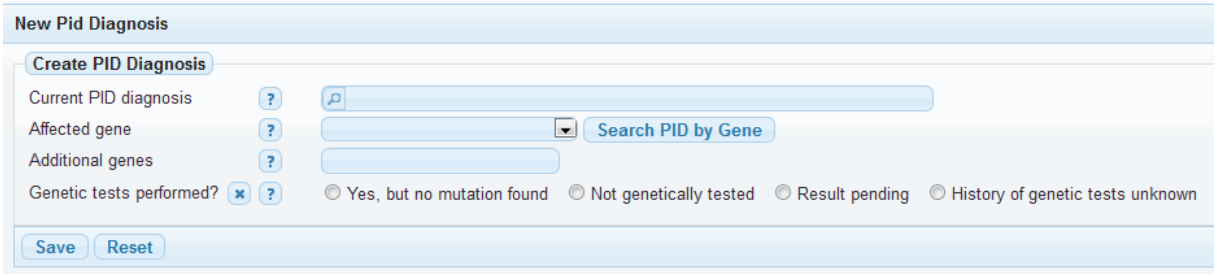
You need to change HSCT and gene therapy only if it was “No” or “Unknown” previously, because the previous selection is already displayed (so “Yes” remains “Yes” and does not have to be selected again at every documentation).

Please also note that all HSCT and gene therapies are only entered once and can be edited at every documentation timepoint.

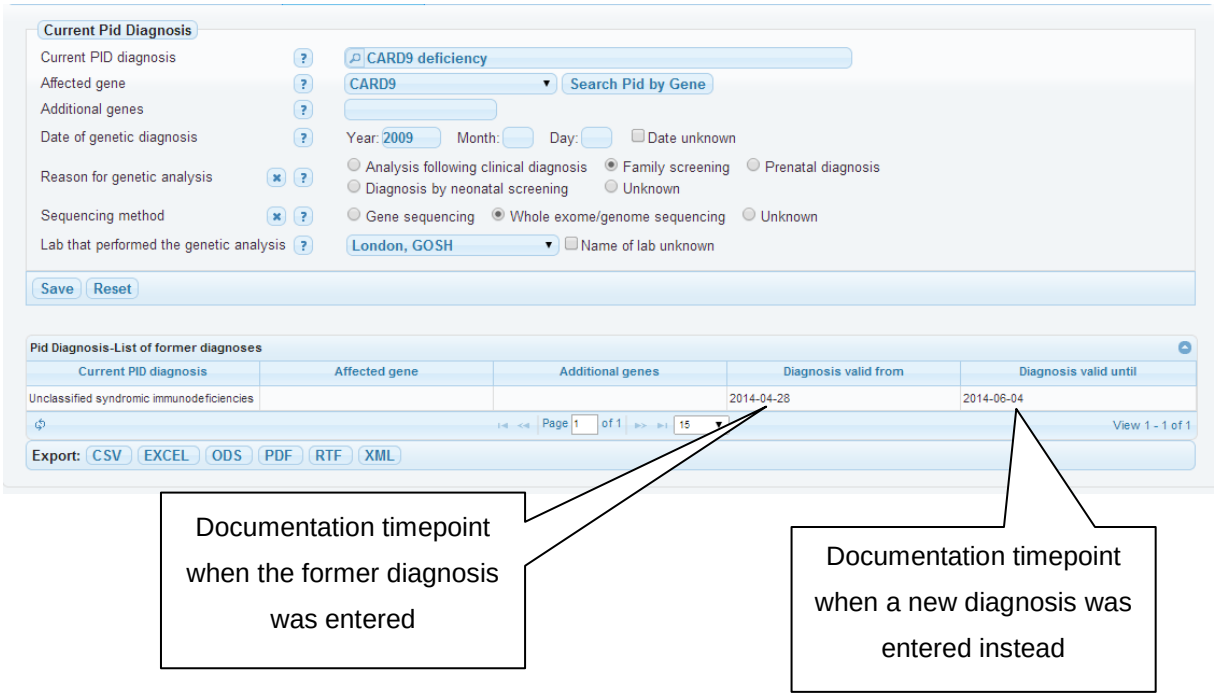
2.8.1 Changing the PID diagnosis

If you have found a genetic mutation in your patient, you can simply enter this into the PID diagnosis. However, if the **clinical diagnosis (“PID Diagnosis”) has changed**, this has to be documented as a new diagnosis entry, in order to preserve the previous data:





Previous diagnosis entries are displayed as a list below the current diagnosis:

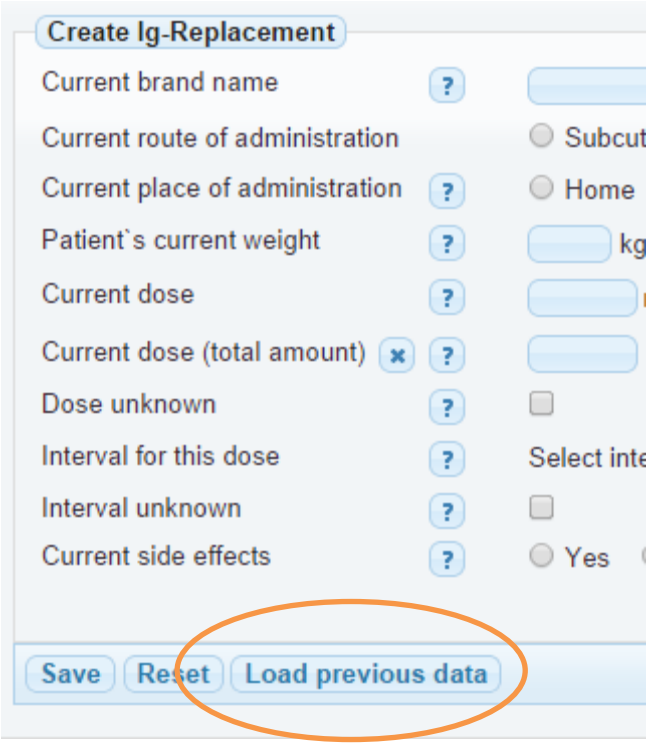


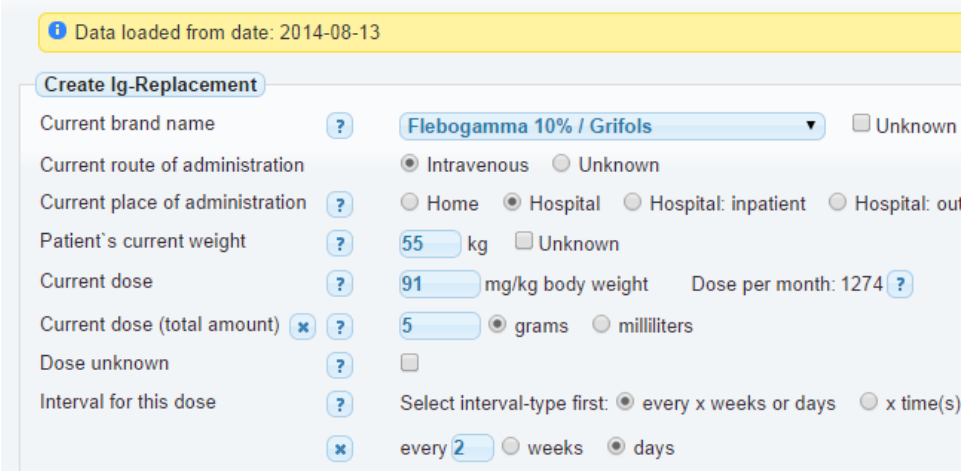
Documentation timepoint when the former diagnosis was entered

Documentation timepoint when a new diagnosis was entered instead

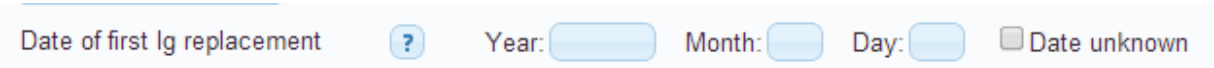
2.8.2 Loading previous data for Ig replacement

In order to minimize the documentation burden, you can load the Ig replacement data from the previous documentation timepoint into the form:





Please also note that the date of the (very) first Ig replacement is only requested at the baseline documentation:



2.9 Personalized version

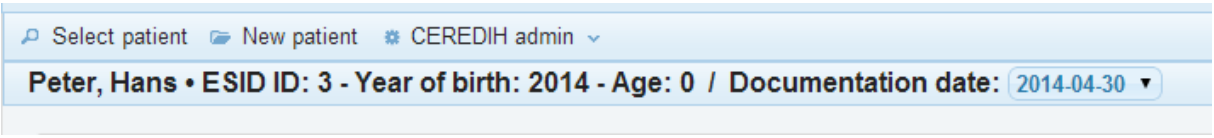
If your centre uses the personalized version, i.e. patient names are included, there are additional features.

2.9.1 Patient list with names

In the patient list, you can display patients' names. You can enable this feature by clicking on "Show patient names":

Please note that if your centre has a large number of patients, it may take up to 15 seconds to load the patients' names because the personal patient data is stored on another server and loaded into the page.
Therefore, you can switch between "Show patient names" and "Hide patient names" as you like. You can also set a default value for this feature in the "Account settings" (2.7).

The patient's name is always displayed on top of the page, independent of the patient list setting:



2.9.1.1 Improving the loading time

In your account settings (see 3.1) you can define that your complete patient list will be saved locally on your computer. Note that first name, last name and complete birth date will NOT be saved, but this caching method still reduces the loading time for the patient list with names considerably, especially if you have more than 1000 patient entries. Please also note that after you have activated this feature, the first

time that you load the patient list, it will not be faster, because then the cache is being generated. On all subsequent loads, the loading time should be faster.

2.9.1.2 Downloading the patient list with names

You can also **download** the patient list with names, using the button below the list:

Currently, only CSV format is available. Depending on your browser, you will need to save and rename the downloaded file to a .csv file. In order to open it with Excel, please use the Excel text import function.
Please make sure to set the CHARSET in the Excel import settings to UTF-8, otherwise special characters like Umlauts will not be displayed correctly.

2.9.1.3 Sorting and filters

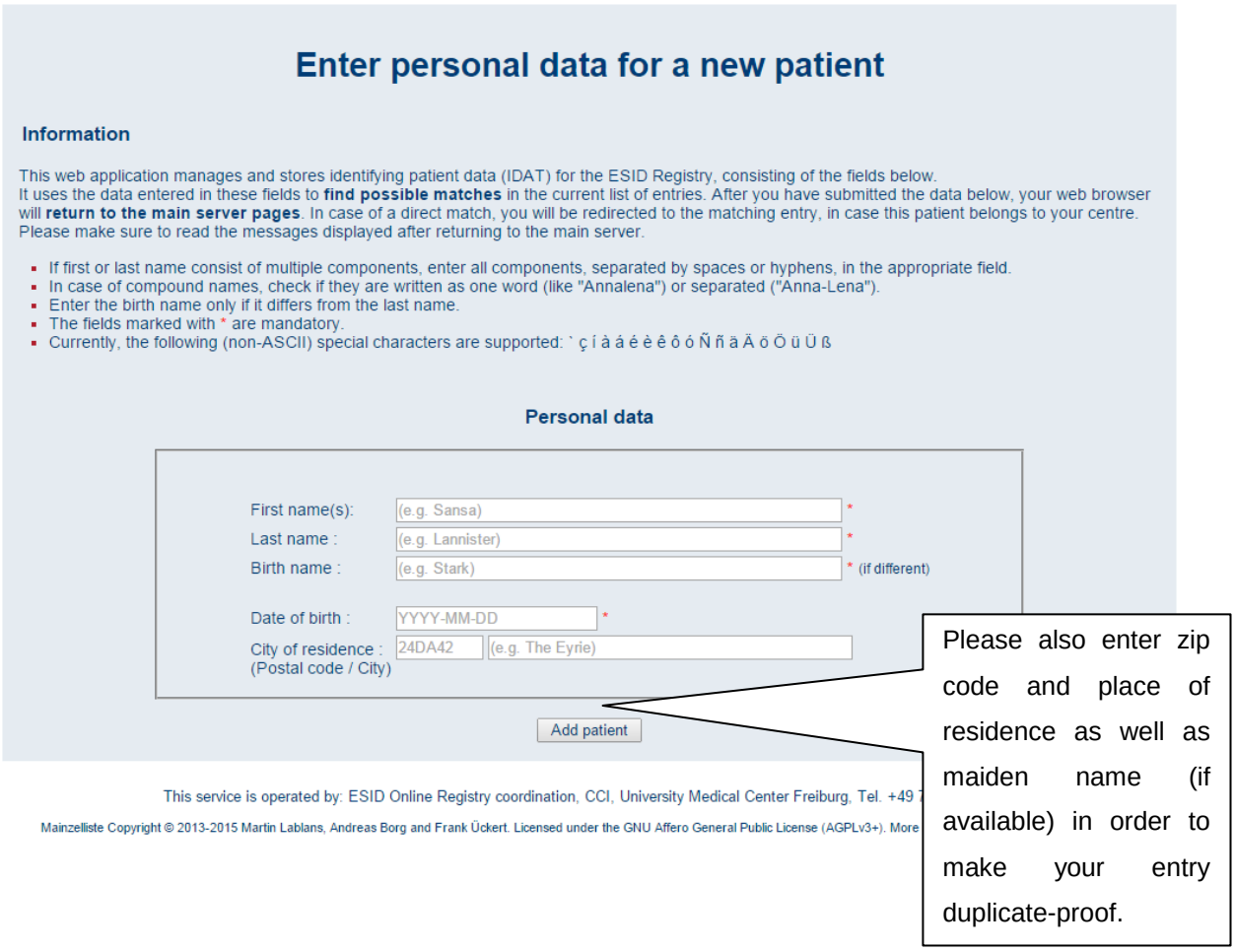
Please note that the sorting and filters that you set and save on the patient list WITH names is NOT visible in the patient list WITHOUT names, and vice versa. This means that in fact, you can save two sets of filters.

2.9.2 Create a new patient

If you create a new patient, the system redirects you to the other server in order to enter the patient name etc. (screenshots next page).
This other server has two purposes:

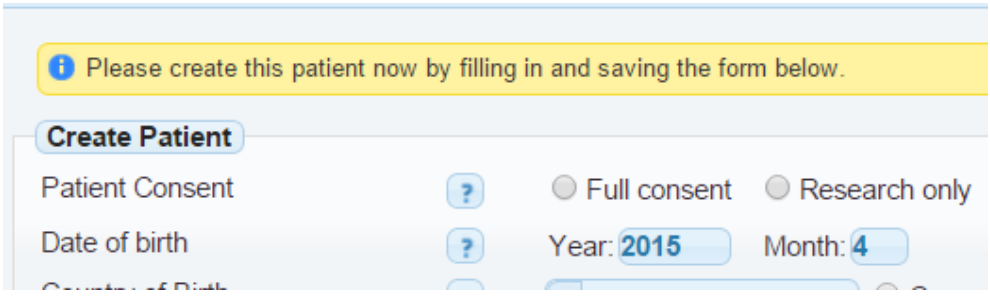
1. It stores the patient name, complete date of birth and place of residence so this data can be loaded into the ESID Registry
2. It performs a so called “record linkage” and thereby prevents reporting of duplicate entries.

For the second task, it is important that you really enter as many details as possible, and not only the patient names.

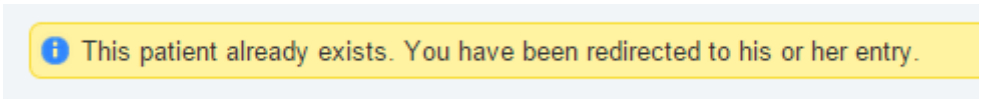

Enter personal data for a new patient
Information
This web application manages and stores identifying patient data (IDAT) for the ESID Registry, consisting of the fields below.
It uses the data entered in these fields to find possible matches in the current list of entries. After you have submitted the data below, your web browser will return to the main server pages. In case of a direct match, you will be redirected to the matching entry, in case this patient belongs to your centre.
Please make sure to read the messages displayed after returning to the main server.
If first or last name consist of multiple components, enter all components, separated by spaces or hyphens, in the appropriate field.
In case of compound names, check if they are written as one word (like "Annalena") or separated ("Anna-Lena").
Enter the birth name only if it differs from the last name.
The fields marked with * are mandatory.
Currently, the following (non-ASCII) special characters are supported: ` ç í à á é è ê ô ó Ñ ñ ä Ä ö Ö ü Ü ß
Personal data
First name(s): (e.g. Sansa) *
Last name : (e.g. Lannister) *
Birth name : (e.g. Stark) * (if different)
Date of birth : YYYY-MM-DD *
City of residence : 24DA42 (e.g. The Eyrie)
(Postal code / City)
Add patient
This service is operated by: ESID Online Registry coordination, CCI, University Medical Center Freiburg, Tel. +49
Mainzelliste Copyright © 2013-2015 Martin Lablans, Andreas Borg and Frank Ückert. Licensed under the GNU Affero General Public License (AGPLv3+). More
Please also enter zip code and place of residence as well as maiden name (if available) in order to make your entry duplicate-proof.

After you have added the data, select “Add patient”, and the system will take you back to the main server.

If the patient has not been registered before, there will be a message in the top row saying:



If the patient exists and has been registered at your centre, the message will say:



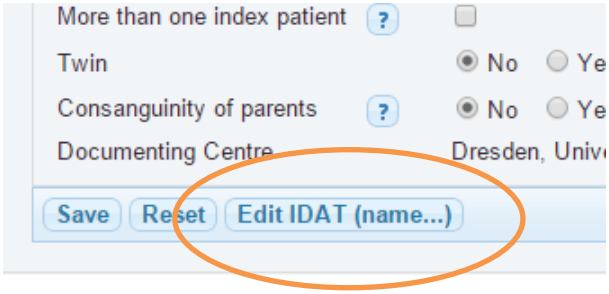
If the entry belongs to **another** centre, you are redirected to your patient list, and there is a **message** that tells you the name of the center that this patient belongs to.

2.9.3 Edit personal information

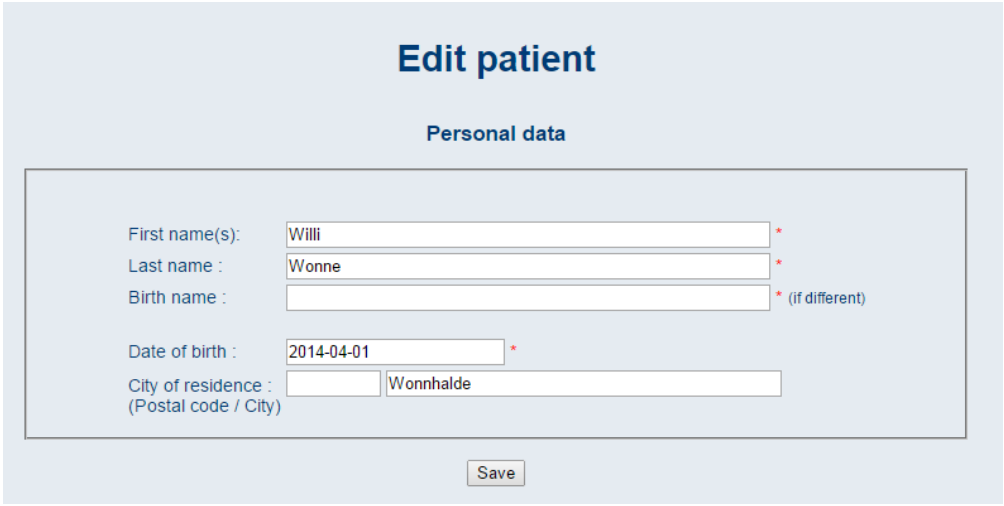
Sometimes, you realize that part of the personal information is incorrect, e.g.

- The patient has married and changed his/her family name
- There is a typo in the entry

You can then edit the personal information by selecting the patient and clicking on the "Edit IDAT[1] (name…)" button in the Patient tab:



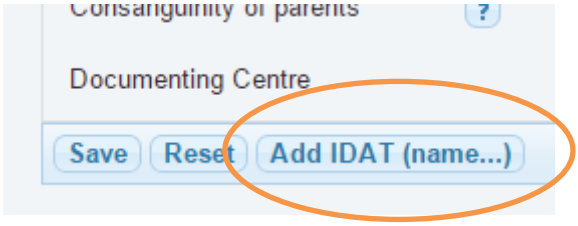
This results in a redirect to the other server, where you can edit all of the data:



When you are finished, "Save" takes you back to the standard view (Patient tabs).

2.9.4 Add personal information to existing patient

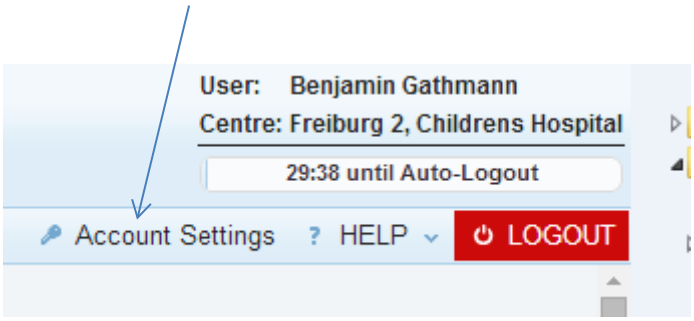
It may happen that some of your patients have been registered without names, e.g. because you recently switched to the personalized version. In these cases, you can add the IDAT by clicking on the following button in the "Patient" tab:

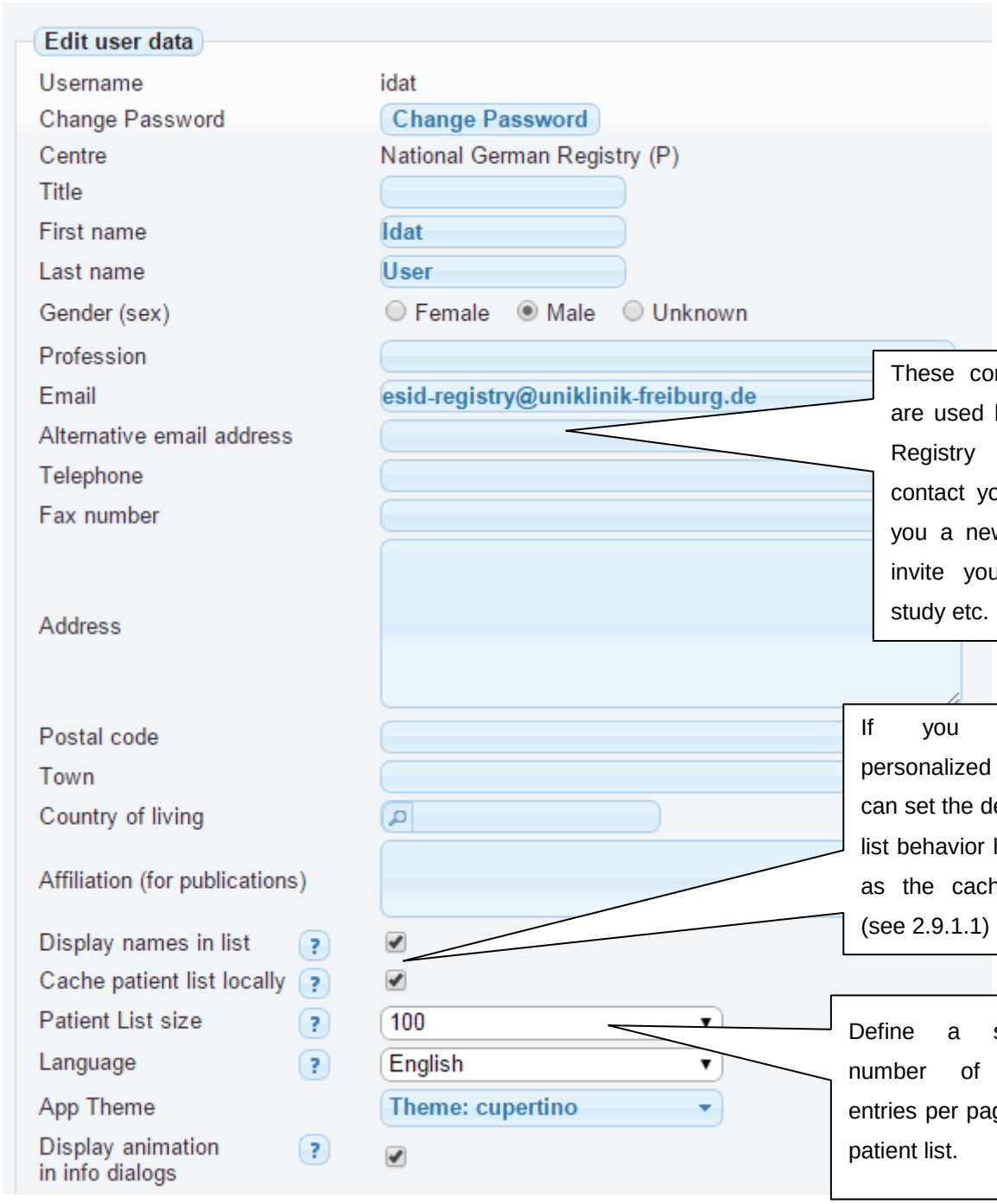


[1] „IDAT" is short for the technical term „identifying data"

2.10 Account settings

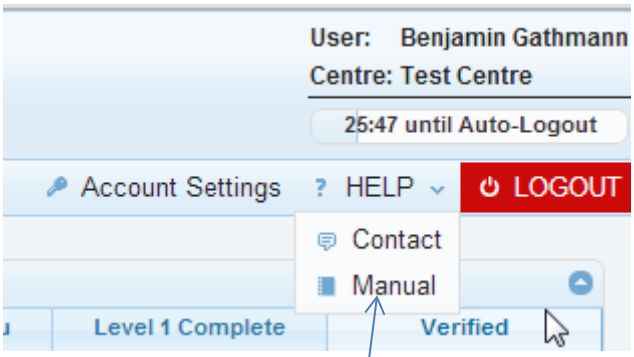
You can edit your account and change your password by clicking the link on the top right of the screen:



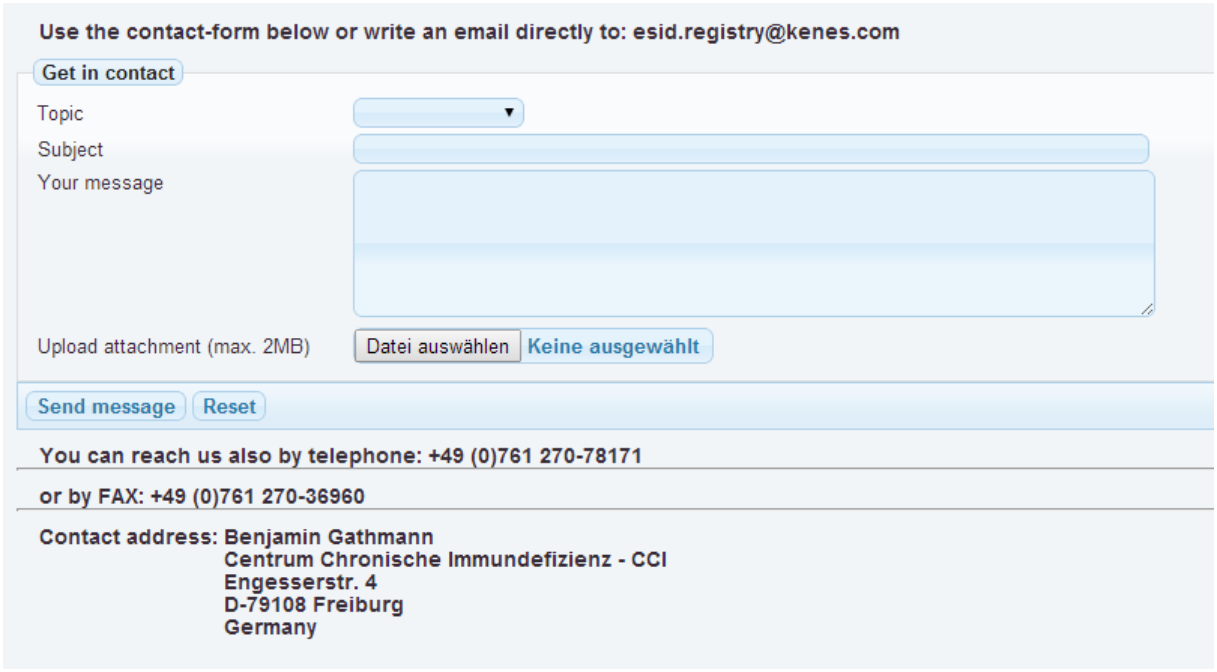


3 Contacting the Registry administrator

For contacting us, you can either send an email or call us (details see below) – or you can use the contact form inside the Registry which is available in the “HELP” menu:



As you can see, this manual itself is also available in the “HELP” section.



<u>Contact details:</u>

University Medical Center Freiburg

Center for Chronic Immunodeficiency (CCI)

Engesser Str. 4

79108 Freiburg; Germany

Tel.: +49-761-270-78171

Email: esid-registry@uniklinik-freiburg.de

4 Registry FAQ

This manual does not answer all possible questions.

A good starting point for questions not answered here are our FAQ which you may find here:

http://esid.org/content/download/13670/386342/file/Registry_FAQ.pdf